KELLY (A.O.J.)

Athetosis

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ATHETOSIS.

BY A. O. J. KELLY, A.M., M.D.

Clinical Assistant in the Neurologic Department, Philadelphia Polyclinic; Pathologist to St. Agnes' Hospital; Assistant to the Medical Dispensary at the Hospital of the University of Pennsylvania.

SYMPTOMATIC only though it usually is, being rarely a disease *sui generis*, of sufficient interest nevertheless is the subject of athetosis. First described by Hammond in 1871, it has since been the subject of repeated investigations and careful study, until to day its symptomatology is as well understood as its pathology is obscure. A report of the following case, presenting, as it does, the symptoms typically, may not be devoid of interest.

The patient, J. M., aged 18 years, male, single, white, a native of the United States, presented himself at the Philadelphia Polyclinic, March 6, 1895. His mother died of right-sided hemiplegia, his father, one sister, and three brothers are living and well. The patient was apparently healthy until he was one year old, when he had a convulsion, which is said to have come on suddenly, and is supposed to have been caused by teething. No acute infectious disease preceded the convulsion, nor is he known to have been particularly ill at this time. Beginning thus, these convulsions, occurring once or twice every two months or thereabouts, gradually increased in frequency until they would occur once a week or oftener. When thirteen years of age, having been under medical treatment more or less all his life, the convulsions ceased, and have not recurred. As he now recalls the attacks, his first sensation was that of a "lightness in the head," followed by winking of the left eye, twitching of the left side of the face, and drawing of the head to the right side. Then came on unconsciousness and convulsion affecting the entire left side of the body. Unconsciousness would continue for from ten minutes to half an hour; then supervened a condition of drowsiness and sleep lasting two hours, or more. On different occasions during the convulsions he bit his tongue, but never had any involuntary voiding of urine that he can remember. He presents himself now to ascertain if the condition of the left side of his body, particularly of his hand, can be improved. The hand is the seat of mobile spasm (Gowers); of movements slow, involuntary, and of some regularity; movements of adduction and abduction, of flexion and hyperextension of the fingers, and of pronation and supination of the hand and forearm, follow each other in a

sequence more or less orderly. (One of the most frequent positions of the hand is repre-

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sented in the accompanying figure). This condition has endured since the patient and his elder sister can remember. The hand is at times for a while quiet. Attempts at voluntary motion always induce the mobile spasms, which become very much exaggerated and markedly inco-ordinate upon emotional or other excitement. Occasionally he can exercise a slight momentary control over the movements of his hand; whether or not these movements persist during sleep he cannot say. He has little or no power in his left hand, and but little in the forearm; good power in the upper arm. The muscles of his arm are well preserved, as are also apparently the muscles of his forearm and hand. The left arm and hand is somewhat smaller and a trifle shorter than the right. The deep and superficial reflexes are slightly exaggerated; there is no disturbance of sensation. The left leg is a little less thick and a trifle shorter than the right; good muscular power; no disturbance of sensation; patellar-tendon reflex slightly exaggerated; no ankle clonus; no spasms of toes, nor can he move his toes at all. The face is probably unaffected, although it occasionally seems, especially when he talks or smiles, as though there were an earlier and some slight overaction of the muscles of the left side. There is slight tremulousness of the tongue on protrusion, but no disturbance of speech. There is no numbness of the affected side, nor any pains at the seats of spasms. The patient sleeps well, has seldom headache, and never vertigo. He has no symptoms referable to his thoracic or abdominal viscera, examination of which is negative.

The diagnosis was made of athetosis following acute cerebral palsy of childhood. Unfortunately, as is often the case, he came to us as a *dernier resort*, and equally unfortunately for him, we could prognosticate no more favorable course than that which the affection had heretofore run. He was given sodium iodid, and ordered to report regularly for electric treatment, in the hope that some benefit might be derived therefrom.

Cases of athetosis are interesting, especially from a diagnostic and pathologic point of view. In the diagnosis we must first distinguish between idiopathic and symptomatic athetosis. The *morbus sui generis* is comparatively unfrequent, and the causes to which it is usually ascribed—*e g.*, traumatism, fright, cold,—are often either absolutely erroneous, or must be accepted with great reserve by an impartial critic. Jacoby, Strümpell, and others speak of a congenital athetosis. Symptomatic athetosis may be attendant upon a variety of nervous disorders. Originally described in connection with epilepsy and severe psychoses, it is as hemiathetosis post-hemiplegica, that we most commonly meet with it; not so often, indeed rarely, following the ordinary hemiplegias, but especially after the acute cerebral hemiplegias of children. Osler, Gowers, Oppenheim, Strümpell, etc., mention this connection, and assert that by far the majority of infantile hemiplegias show sometime or other during their course more or less well developed athetosic movements.

The only tremor with which athetosis is likely to be confounded is that of chorea, and with care even this should not happen. The athetosic movements are to be distinguished from those of chorea by their localization and by their character. By their localization, in that they are usually confined to the peripheral parts of the extremities, the hands and feet, the choreic spasms being not thus limited. Marked athetosic implication of the face is very uncommon. Indeed, if the face alone be affected, the disorder is almost certainly choreic. In character, the slow, more or less regular and monotonous movements of athetosis serve to distinguish it from the quick, eminently irregular, and purposeless movements of chorea. A relationship be-

tween the two can, however, not be denied. Leube reports a case of idiopathic athetosis which gradually assumed the character of a typical chorea. Bernhardt cites a case of post-hemiplegic chorea, in which the movements became athetosic. In this connection, Leube says that post-hemiplegic chorea, and post-hemiplegic athetosis are identical. Oppenheim mentions that he saw cases in which the clinical picture of the upper extremity was that of chorea, while that of the lower extremity was athetosis.

The pathology of the affection has not, as yet, been satisfactorily explained. Within late years there has been a seeming tendency among some to accept the view that a lesion anywhere in the course of the fibers of the pyramidal tract may be followed by the production of athetosis. Strümpell, however, says, that concerning the nature of athetosis, concerning the region where the irritation occurs, and the manner in which it develops, nothing is as yet known. He is disposed to believe that it is always due to some cerebral, perhaps cortical, disturbance. But he mentions a personal observation of his,—a case of well-developed athetosic movements of the arm and hand, which developed idiopathically in an aged woman, and in which the post-mortem examination of the brain gave absolutely negative results. Those believing that disease affecting any part of the pyramidal tract may cause athetosis, base their assertions on the great variety in the localization of the lesions found post mortem. Démange has reported a case proving conclusively that disease limited to the cortex may cause athetosis. Kahler and Peck have endeavored to demonstrate that all disturbances of motion following hemiplegia are due to disease of the pyramidal tract between the optic thalmus and the lenticular nucleus, the various sorts of disorder of motion to be explained upon the supposition of destruction, partial or complete, or simple irritation of the fibers. And many cases have been reported in which there were post-mortem evidences of disease in this region. In some cases it has been disease of the thalmus; again, disease of the lenticular nucleus; or the fibers themselves of the pyramidal tract have been implicated. The two former would have affected the latter indirectly.

Not the least plausible argument is that offered by Gowers, who suggests that the athetosis is due rather to the quality of the lesion than to its site. He bases his supposition upon two important factors in the etiology: firstly, that the disorder is much more frequent after cerebral softening from vascular occlusion than after cerebral hemorrhage; secondly, that it follows the hemiplegias of childhood much more frequently than it does those of adult life. "The probable significance of the first fact is that in softening slight damage to the cerebral tissue is more extensive than the actual destruction, and the spontaneous spasm must be referred to the overaction of gray matter, which is in a state of altered nutrition and function. Hence we can understand the occurrence of this symptom from a lesion which involves extensive slight damage. The significance of the second fact—the frequency with which the condition follows infantile hemiplegia—is probably the greater facility with which the growing and developing nerve cells recover, and their greater susceptibility to disorder of function where their development is perverted." (Gowers.) The lesion is thus one of quality—impairment of the nutrition of growing motor nerve cells. But the uncertainty is well expressed: "We cannot yet give any trustworthy explanation of the mechanism of the spasm in the limbs which so constantly accompanies the hemiplegia. It seems to follow lesions of various kinds, degrees and seat, in the cortex as well as the central ganglia. The far greater frequency (almost constancy) of the symptom after a lesion in early life makes it probable that it

is in some way due to the disordered action of centers that remain, and not to the direct effect of the disease itself." (Gowers.) Regarding idiopathic athetosis, or athetosis occurring without hemiplegia, the supposition is that it is caused by a lesion not sufficiently severe nor extensive enough to produce a paralysis, the lesion being more irritative than paretic producing.

Not without interest in the case reported, and in many similar ones, is the occurrence of convulsions, which in all essential particulars resemble true epilepsy, and in their mode of origin simulate focal or cortical epilepsy. These are the convulsions occurring during the course of a vast majority of the cases of infantile hemiplegia, and constitute not an inconsiderable proportion of the cases of epilepsy as we ordinarily meet with them. They frequently, as in the case reported, begin in some one particular locality, and are usually confined to the paralyzed side. At times, apparently as in our case, the convulsions may commence at the time of the occurrence of the hemiplegia, and continue, recurring at irregular intervals; in the majority of cases, however, there is usually quite an interval between the attack of hemiplegia and the development of the epilepsy. It is not uncommon to hear of the hemiplegia having been acquired in early childhood, the development of the epilepsy being postponed until the so-called epileptogenic period of puberty. Not to pass unnoticed in the case reported is the sudden cessation of the convulsions when the patient was thirteen years of age, since which time he has been entirely free from them. He still occasionally takes some of the medicine—probably bromides—which are supposed to have stopped them.

But little is to be said regarding treatment. The nature of the lesions producing the affection, so far as they are known, is such as to almost exclude the possibility of any beneficial influence being exerted by medicines. Electricity is of but slight service. Some cases of improvement have been reported in which galvanism, bromides, iodides, etc., were used. But unfortunately we can promise little.

In conclusion, to Dr. Chas. K. Mills, I desire to express my appreciation of his uniform kindness to me, both in permitting me to report this case, and for the photograph.